

.616851

A THESIS

SUBMITTED FOR THE DEGREE OF M.D.,

EDINBURGH UNIVERSITY.

A STUDY of SYDENHAM'S CHOREA

by

GEORGE HAROLD LOWE,

M.B., Ch.B., 1907.

M.D. 1912.



INTRODUCTION.

I have lately had under my care a series of 46 cases of Sydenham's Chorea of varying intensity. It is my purpose in this Thesis to describe the disease in its Etiology, Pathology, Symptoms, and Treatment, with some reference to these.

Chorea is the disease popularly but erroneously known as St Vitus' Dance. Other synonyms are Chorea Minor, Chorea Vulgaris, Chorea Anglorum.

The derivation of the word is from the Greek Xopeia which means Dancing and the name was unfortunately given to the disease under consideration by Sydenham who described the condition in 1686 and 1693. The reason for this unfortunate name being given to the condition is that Sydenham mistook it for a Dancing Mania which was prevalent in some parts of Germany in the fourteenth and fifteenth centuries. This was a religious mania characterised by wild delirium and dancing till the person afflicted dropped from sheer exhaustion. It was of a hysterical nature and spread by imitation.

St Vitus was a Sicilian who suffered martyrdom at the persecution of Christians under Diocletian. The legend arose that St Vitus had prayed that Heaven would protect from the Dancing Mania all who should observe his commemoration day. Sufferers from the mania visited the Shrine of St Vitus where the priests performed a religious ceremony to cure them /

them. Various shrines were visited in this way in different parts and the disease also was known as St John's, St Anthony's etc. Dance.

St Vitus' Dance was at first considered as belonging to the moral and spiritual man and not to his body and the condition was handed over for treatment by the physicians to the priests. Paracelsus was the first to bring the disease under medical notice. He named it Chorea Sancti Viti.

Having briefly described the Dancing Mania I shall now give some description of the Disease which is the subject of this Thesis. Sydenham defined it as "a Kind of convulsion attacking boys and girls from the tenth year to puberty" and really the description could not very well be bettered.

Brissaud's definition was "Purposeless and seemingly causeless involuntary movements occurring both during activity and rest and therefore illogical and maladroit." Sydenham after defining Chorea goes on to say "It first shews itself by limping and unsteadiness in one of the legs which the patient drags. The hand cannot be kept steady for a moment. It passes from one position to another by a convulsive movement however much the patient may strive to the contrary. Before he can raise a cup to his lips he makes as many gesticulations as a mountebank, since he does not move it in a straight line, but has his hand /

hand drawn aside by spasms until by some good fortune he brings it at last to his mouth. He then gulps it off at once so suddenly and so greedily as to look as if he were trying to amuse the lookers on."

Sydenham does not enter into the etiology of the disease but his treatment is quaint. It was as follows :-

Bleed the arm to eight ounces more or less according to the age. Next day give half (more or less as the age of the patient requires it) of the common potion. In the evening the following should be taken :-

Black cherry water.

Aqua Epileptica Langii

Venise treacle

Liquid laudanum.

Make into a draught.

Repeat the cathartic every other day, three times and the paregoric on the same night. Blood must again be drawn the next day and the catharsis repeated, and so bleeding and purging must alternate till the third or fourth time provided there be sufficient time between the alternative evacuations to ensure the patient against danger. To guard against a relapse, bleed and purge for a few days next year at the same time or a little earlier.

Since /

Since Chorea was first established by Sydenham as a separate clinical entity, the disease has been described by many observers. Between 1693 and 1800 many contributions were made to the literature. Reference to these was made in a paper by Berni~~y~~ in 1810 in which year an important addition was made by Bouteille in France. In that country the chief contributions have since been made by Trousseau, See, Roger and Charcot ; in Germany by Romberg, Steiner, and Von. Ziemssen ; in America by Weir Mitchell, Jacobi, Putnam, Osler and others ; but the most important have come from this country - from Bright, Todd, Wilks, Fagge, Hughlings Jackson~~s~~, Gowers, Poynton, and many others. All their definitions may be summed up in describing the condition as a nervous disease usually affecting children and characterised by involuntary, irregular, spasmodic, muscular contractions, a variable degree of muscular weakness associated with some inco-ordination of voluntary movements, and by psychical disturbance usually partaking of the nature of hebetude and mental weakness.

The following is an extract from Wilk's Diseases of the Nervous System, 1883 :- "I doubt whether Chorea is due to any special disease of the spinal cord or other part of the nervous system but is not rather, like epilepsy, due to a disturbance of the whole /

whole of the centres. That the brain is affected is shewn by the occasional maniacal excitement and the more frequent tendency to imbecility. Just as in epilepsy you might imagine a sudden disruption or discharge of nervous force exciting the ganglia below and temporarily suspending the action of the cerebral hemisphere in which the explosion took place, so, in Chorea, you might regard the irritation as more continuous, rendering the movements therefore constant. Hence when any extra work is put on the cineritious matter of the hemispheres, as when volition comes into play, the movements are increased. The condition is one in which the nervous centres have become irritable and lost their power, and the will is incapable of directing their action."

By some Chorea is claimed as a functional brain disorder affecting the motor centres, an instability of the motor nerve cells. By others it is claimed to be organic in origin, either depending on some gross lesion of the brain or on some bacterial or toxic agent. When discussing the etiology and pathology of the disease I shall endeavour to set forth the arguments for both theories assisted by my own experience.

Chorea is much more common in the lower or hospital classes than in the better class of private practice /

practice. The reason may be that the latter are better cared for and nourished. In the Report of the Collective Investigation Committee of the British Medical Association 72.27 per cent of cases were in the lower classes.

In many instances there is a hereditary tendency. One may find that there has been Chorea in a previous generation. More than one member of a family may be affected. It is more common, however, to find that the patient is the neurotic child of a neurotic father or mother or both, than that there is a direct hereditary tendency to Chorea. One or both parents may be the subject of epilepsy or there may be a taint of insanity in the family history.

With this short introduction I shall proceed to discuss the various aspects of the disease.

ETIOLOGY.

Under this heading I propose to group the various factors, predisposing and exciting, which are commonly accepted as having some influence over the incidence of the disease. These are 1, Age. Chorea is almost essentially a disease of later childhood and puberty. Cases do occur in adults, but they are rare and are probably not primary attacks. The usual age is between 6 and 15 years, Of 46 cases under my own care the youngest was aged 7 years and the oldest 23 years. The former was a primary attack while the latter was a recurrence. The average age was 13 years.

Under : The disease is rare beyond the age of 5 years although a few cases, said to be undoubtedly congenital, have been reported. In the Report of the Collective Investigation Committee of the British Medical Association 34 per cent were between 5 and 10 years, 43 per cent between 10 and 15 years, and 16 per cent between 15 and 20 years, but it was not stated how many of the last were primary attacks. Of 522 cases analysed by Osler, 33 were in the first hemidecade, 228 in the second hemidecade, 212 in the third hemidecade, 62 in the fourth hemidecade.

The disease attacks girls rather earlier in life than boys.

2. Sex. There is no doubt but that Chorea is much more common in female children than in males.

This /

This has been the experience of every observer and a few figures may be given illustrative of the fact.

Of the 46 cases under my own care, 36 (or 78 per cent) were female, while 10 (or 22 per cent) were male. My experience therefore is that females exceed males in the proportion of nearly 4 to 1.

Of 554 cases in the Philadelphia Infirmary for Diseases of the Nervous System analysed by Osler, 71 per cent were female and 29 per cent male. According to Osler the percentage of females increases after puberty. This I can corroborate as only 1 of my male cases was over 15 years of age, while 9 females exceeded that age.

Of 808 cases in John Hopkins Hospital, 71.2 per cent were female.

Gowers considers the relative proportion to be 3 girls to 1 boy.

The preponderance of female cases over males may be explained by the fact that Chorea most commonly occurs in neurotic highly strung children, the majority of whom are female.

3. Race, Geographical Distribution and Climate.

Chorea is widely found in the United Kingdom and in America. It is essentially a disease of temperate climates. In this country more cases are encountered in the towns than in the country districts, but it cannot /

cannot be said that proportionately to the population Chorea is more common in towns.

In America the disease has been found to be rare among negroes and Indians, and practically universally an affection attacking the white population. It occurs in half breeds but Osler never traced it in full blooded Indians. Only 25 of the cases in John Hopkin's Hospital were negroes.

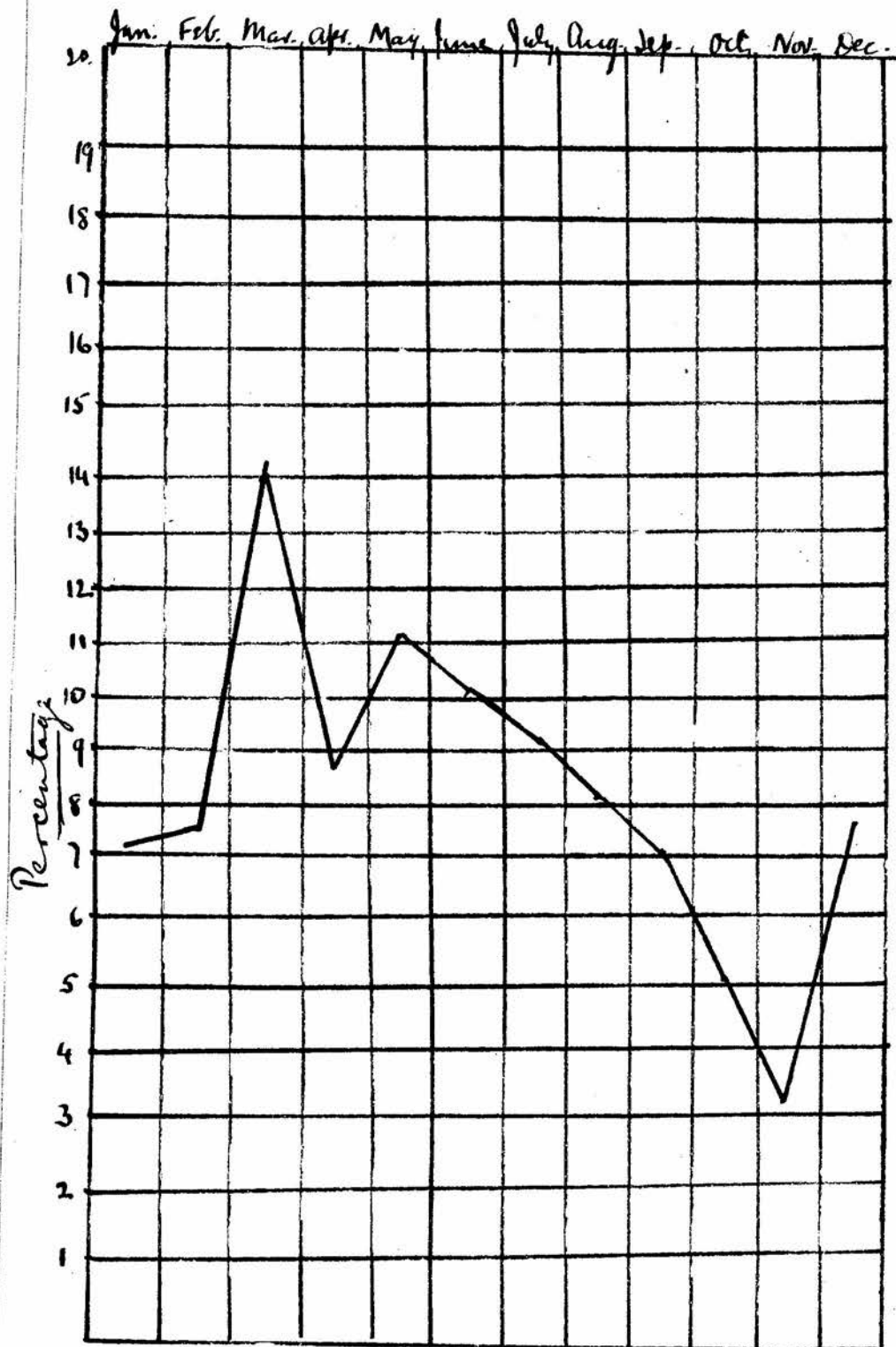
Chorea is also common on the Continent of Europe. From Germany and France many valuable contributions to the literature of the subject have emanated.

4. Season of Year.

Morris Lewis of Philadelphia has established a distinct relation between the incidence of Chorea and the season of the year. He found that in an analysis of 717 separate attacks the highest percentage occurred in the spring months particularly in March and the lowest in November (see table appended) He concluded that cases are most numerous when the mean relative humidity is excessive and the barometric pressure low.

In this country the season of the year does not seem to have much effect. This is not to be wondered at when one considers the vagaries of the British climate. Sir Wm. Gowers found that of 100 cases, 33 occurred from January to March, 25 from April to June, 20 from July to September and 22 from October to December. Dr Emmett Holt of New York found that most cases occurred in May.

Table illustrative of Seasonal Incidence (Lewis)



5. Rheumatism.

Regarding the relationship between Rheumatism and Chorea there has existed in time past a diversity of opinion, but at the present time the fact that both acute Rheumatism and Chorea are dependent on the same causal factor is almost universally recognised.

Some writers regard Chorea as a late manifestation of Rheumatic Fever. My own experience has been that in a number of cases there has been a distinct history of Rheumatism, in one or other of its forms, including the "Growing Pains" of childhood, or a rheumatic family history.

The association attracted the attention of the early writers Hughes, G. See, and H. Roger who in 1866 regarded Rheumatic Fever, Chorea, and Endocarditis, as different names applied to one and the same condition.

In this country the first recognition of the association was made by the Physicians at Guy's Hospital, London. According to Bright Rheumatism was regarded as the cause of Chorea in 1802. According to Babington the credit of the discovery belongs to Addison.

The Rheumatic causation of Chorea has been more accepted by the French than by the Germans, with the exception of Mayer who ascribes 80 per cent of his /

his cases to Rheumatism. Osler gives statistics of two series of cases of Chorea with reference to Rheumatism. In one series of 554 cases, 15.5 per cent had a family history of Rheumatism and in 15.8 there was a history of articular swelling. In another series of 175 cases 18.24 per cent had a family history of Rheumatism.

Here regards Rheumatism as only a contingent cause comparable to other infectious diseases.

In those cases where there is undoubted connection between the two conditions, as when they are co-existent it may be difficult to say whether the rheumatism preceded the Chorea or vice versa. Statistics have been collected by various observers bearing both upon the actual occurrence of rheumatism in the individual prior or subsequent to the attack of Chorea and upon a personal or family history of arthritic affection. The largest recorded proportion in the former category is 50 per cent. In the latter Tylden found it as high as 72 per cent. The Collective Investigation Committee of the British Medical Association found that rheumatism preceded Chorea in 26 per cent ; in 32 per cent the rheumatism accompanied the Chorea or occurred subsequently. The latter percentage rose to 46 if patients subject to vague pains were included. Starr found a rheumatic history in 18 per cent of

385 cases ; C. W. Townsend 21 per cent of 148 cases ; Crandall, 57 per cent of 111 cases. In 41 of Crandall's cases rheumatism preceded Chorea ; in 13 the two were coincident ; in 9 rheumatism occurred subsequently to the attack of Chorea, usually within 3 months. According to Whipham in less than 2 per cent of cases does Chorea precede Rheumatism. Batten found that 11.3 per cent of patients with Chorea acquired rheumatism in 3 years and 20 per cent in 6 years.

327 cases under Morley Fletcher shew that Chorea was preceded by Rheumatism in 26 per cent. Sir S. Mackenzie gives the figure at 29 per cent and Sturges at 26.5 per cent.

Dickinson found antecedent rheumatism in 26.75 per cent and coincident in 7 per cent.

Peacock found antecedent rheumatism in 28 per cent and coincident in 7.66 per cent.

Owen found antecedent rheumatism in 26 per cent and coincident in 8 per cent.

Ogle found coincident rheumatism in 10 per cent.

Woolenberg found antecedent rheumatism in 38 out of 50 cases, and coincident rheumatism in 3.9 per cent of 51 cases.

Steiner found coincident rheumatism in 1.6 per cent of 252 cases.

Pryor found that in 92 cases with antecedent or coincident rheumatism 5.4 per cent had endocarditis.

Osler /

Osler found endocarditis in 54 out of 110 cases and 40.7 per cent of these had articular pains.

Hughes found that 86 out of 104 cases had rheumatism or endocarditis.

See found that 61 out of 128 cases had rheumatism or endocarditis.

Burton Brown found that 89 out of 104 cases had antecedent rheumatism.

Chapin's experience was that 58 per cent of cases had a history of rheumatism and that 13 per cent had organic heart disease.

Piper found that 46 per cent of cases had either antecedent or coincident rheumatism.

Herringham found articular rheumatism in 26.2 per cent and rheumatic pains in 20 per cent of cases.

Meyer found rheumatism in 9 per cent and heart lesion in 10 per cent of cases.

Suchs found that of 184 cases 20 had antecedent rheumatism.

Ziemssen, Mackenzie and Gowers find antecedent or coincident rheumatism in 24-26 per cent.

Starr found that of 2476 cases antecedent rheumatism occurred in 26 per cent and coincident rheumatism in 20.3 per cent.

From these figures one is justified in inferring a close association between rheumatism and Chorea in about 25 per cent of cases.

The /

The percentage of cases preceded by rheumatism varies with the age of the patient. Sir Wm. Gowers had only one case under the age of nine years with a previous history of rheumatism.

Poynton and Paine have succeeded in isolating from the cerebro-spinal fluid of patients with Chorea a diplococcus identical with the *Diplococcus Rheumaticus*. This they have cultivated and inoculated rabbits with it. The result was that Choreiform twitching of muscles was produced as well as many commonly accepted rheumatic lesions such as endocarditis, pericarditis, myocarditis, cardiac dilatation, arthritis, subcutaneous nodules, and teno-synovitis. These experiments have been confirmed by Beattie of Sheffield. Poynton and Paine have found this diplococcus in the *pia mater* and brain of patients, who died of Chorea and of rabbits experimentally inoculated with the diplococcus.

Poynton and Holmes have isolated the diplococcus from the blood in 6 cases of ordinary Chorea and of Chorea gravidarum, but in at least one of these *B. Coli*. was found in the blood at autopsy.

With the same organism Vernon Shaw produced cardiac lesions and arthritis in monkeys.

Further reference to these experiments will be found under "Bacteriology."

6. Endocarditis :-

Some writers believe that Chorea is due to Endocarditis which may or may not be rheumatic in nature. Their theory is that irregular movements are due to tiny particles of vegetations becoming detached from the valve and causing embolism of the smaller cerebral arteries. This is commonly known as the "Embolic Theory" and it was first put forward by Kirkes. He found a strong supporter in Hughlings Jackson, who believed that the seat of the lesions was the Corpus Striatum. The Argument advanced against him was that such embolism would cause loss of power or complete paralysis of the muscles. Jackson insisted that this argument favoured the embolic theory rather than opposed it since Chorea was known to be often of a hemiplegic type. He also argued in favour of embolism, the fact that patients lying in bed with endocarditis frequently develop Chorea. The great consensus of opinion at the present time is that the embolic theory, while, no doubt correct in some cases, does not explain the etiology of most cases of Chorea. Chorea occurs without endocarditis, at least in so far as can be found by physical examination. But in several cases where no endocarditis could be determined during life, it has been found at autopsy.

Again /

Again, endocarditis may be present and a histological examination of the brain reveal no emboli to account for the Chorea.

Rosenthal and Money produced Chorea in animals by injecting fine particles into the carotid arteries, thus advancing another argument in favour of embolism.

7. Infectious Diseases.

Chorea is regarded by some as due to infectious diseases, the toxins of which set up the characteristic muscular contractions. In favour of this it is urged that it is often impossible to refer Chorea to endocarditis or endocarditis to rheumatism, and that both originate in some common cause, some infectious agent capable of setting up arthritic disturbance in predisposed people. Such disturbance has been known in Scarlatina, Puerperal fever, pneumonia, rheumatism, and Gonorrhoea. This certainly suggests some association with an infective process.

Against this view, is brought forward the fact that Chorea is very often due to some psychical disturbance., such as fright, or some sudden emotion but too much reliance cannot be placed upon this.

The /

The paresis of Chorea may be analagous to that associated with typhoid fever and other infectious diseases.

Scarlatina may be directly antecedent to Chorea.

Sturges declares that Whooping cough occurs more frequently in Choreic than in other children, but this is denied by Osler, who says that with the exception of Rheumatic Fever, there is no intimate relationship between Chorea and the acute diseases of children.

Infectious diseases are held by some to be rather indirect exciting causes of the attack. In 533 cases of Scarlet Fever, only three developed Chorea (Carslaw). Osler got a previous history of Scarlet Fever in 141 out of his series of 554 cases of Chorea.

Chorea may arise directly after or during an attack of Pyaemia or Gonorrhoea.

It is confidently claimed that organisms may gain entrance to the blood stream through erosions in the mucous membrane of the nasopharynx of children and set up Chorea, (Straton). The disease has occurred the secondary stage of Syphilis and has been cured by anti-syphilitic treatment (Harrison) Mettler).

It has also been associated with Malaria but there is no proof that the condition is more common where Malaria is prevalent.

With the periodic onset /

onset of the fever, the spasms cease. According to Maragliano Chorea may be due to the action on the nervous system of the toxins of staphylococci.

8. Intoxications.

Demme reports a case where Chorea appeared after dressing with Iodoform a fistula connected with caries of the cervical vertebrae. The Choreic movements ceased when the Iodoform was discontinued, and reappeared on the resumption of the treatment.

9. Anaemia is insisted on by some as a factor in the causation of Chorea. It may be so, but it is more likely to be the coincidence that both diseases occur about the same age and usually in females.

Litten had two cases of Chorea developing during the course of pernicious anaemia in both of which recent endocarditis was found at autopsy.

10. Pregnancy.

A patient the subject of Chorea may become pregnant, but usually the Chorea develops during pregnancy. It may develop post partum.

Buist of Dundee arranged the cases recorded up to 1895. Of 226 cases in 6 Chorea preceded pregnancy. In 105 it developed during pregnancy, 31 of which were /

were recurrent. In 16, Chorea was post partum. Forty-five cases were fatal.

In most cases no other cause can be found except the pregnancy although as already stated Poynton and Holmes have isolated D. Rheumatism^{cus} from the blood in Chorea Gravidarum. A pregnant woman is more likely to develop Chorea if she has previously suffered from it. It is more frequent in primipara than in multipara and in connection with this the question arises, Is Chorea Gravidarum due to emotion at discovery of pregnancy? Sir S. Wilks answers in the affirmative and observes that most cases of Chorea Gravidarum are in unmarried girls.

A woman who suffers from Chorea during her first pregnancy may have a recurrence in her second, but more rarely in subsequent pregnancies (Risien Russell). Fare records a case of Chorea in five pregnancies. She may escape during first and develop the affection in a subsequent pregnancy. Chorea Gravidarum scarcely ever occurs for the first time after the age of 25 years.

French and Hicks record three cases aged 30 years.

McCarthy has recorded a case of Chorea in Pseudo-pregnancy.

11. Psychical Causes, such as fright, is often considered a cause, but it is often difficult to trace a connection between the alleged fright and the incidence of the disease. A history of a fright is to be taken very guardedly. It is usually said to have taken place some days before the Choreic movements were noticed. Occasinnally, however, the Chorea develops on the same day on which the fright occurred, leaving no reasonable doubt as to the relationship of cause and effect (Romberg, Von.Ziemsson, Trousseau). But it is rare for Chorea to follow a fright without an interval although Sir Wm. Gowers reports two cases where such a thing really did happen. Also it is rare for the interval to exceed a week.

Other psychical causes are grief, worry, sudden shock, injuries, accidents, surgical operations. In the last named, the associated emotion is more likely to be the causal element. Children the subjects of Chorea are usually very nervous and high strung, and their emotions are very easily excited. They are frequently hysterical, and those cases which are said to be due to imitation are more probably of a hysterical nature. Fagge and Addison accepted imitation as an undoubted cause but the general opinion now is opposed to this.

The strain of education is certainly responsible for Chorea, particularly in highly strung girls about puberty. These girls are often exceedingly bright and intelligent /

intelligent and are urged to work hard for prizes and other honours at school. The result is that the unstable nervous system gives way, and Chorea develops.

12. Ocular Defects were claimed by Stevens as a cause, but in the majority of cases Chorea continues after these defects have been remedied. De Schweinitz shewed that ocular defects are no more common in Choreic than in non-Choreic children, but he admitted that Chorea improves after errors of refraction have been corrected.

PATHOLOGY.

The opportunities for studying the pathology of Chorea have necessarily been comparatively small owing to the very low mortality of the disease. Death occurring in patients suffering from Chorea is usually due to some intercurrent disease with the result that at autopsy and even in a minute examination of the tissues it is often impossible to differentiate the lesions of the intercurrent disease from those which might be due to the Chorea.

With the ever increasing perfection of the microscope, however, advances have been made in the study of the pathology of the disease and various observers have put on record the result of their research. But even at the present time our knowledge of the pathology of Chorea is very indefinite and uncertain and we cannot say that any of the conditions to be described below are really pathognomonic of the disease.

The result of naked eye examination of the Central Nervous System has usually been negative but several conditions have been described. According to Raymond the most common is Hyperaemia, secondly, softening consequent on embolism of cerebral vessels, and thirdly Chronic Encephalitis. Hyperaemia one can understand and readily reconcile with the tremendous expenditure of nervous and muscular energy. Softening /

Softening due to cerebral embolism is what one would naturally expect from such a lesion. To my mind it is impossible to state that any one of these can be definitely associated with Chorea itself.

Dana in a series of 16 autopsies found hyperaemia, punctate haemorrhages, perivascular exudation, foci of softening, and emboli. All these changes were most marked in the basal ganglia. Similar lesions were found by Delcourt and Sand.

Orzechowski^w found foci of degeneration in the motor area but regarded them as septicaemia. He was of opinion that septicaemia was the cause of death.

Occasionally gross lesions are met with, e.g. softening of a large portion of the brain due to embolism of a large cerebral artery. Considerable and fatal cerebral haemorrhage has been found.

Microscopically, examination of the motor of the cerebral cortex has given different results. F.C. Turner in 5 cases found some of the large pyramidal cells swollen and cloudy. Dana found hyaline degeneration in pyramidal cells but his case was complicated by chronic leptomeningitis. Meym^ert found swelling and hyaline degeneration in the cells of the central ganglia and Elischer found similar changes in the Claustrum and the Island of Reil.

Dickinson /

Dickinson found changes in the Corpus Striatum and medulla in several cases. These were erosions and haemorrhages visible to the naked eye and shewing evidence of long-standing by atrophy of the tissue and the presence of haematoidin crystals. These changes were associated with dilatation of the smaller vessels of the medulla. He concluded that Chorea is caused by hyperaemia due to two causes (1) Rheumatism ; (2) Mechanical or mental shock acting more directly on the nervous system. But, generally, speaking, the changes described in the pons, medulla, and spinal cord, are no more characteristic than those of the cerebrum. They have chiefly been hyperaemia, perivascular exudation, and punctate haemorrhages. Steiner has recorded a case of haemorrhage into the central canal of the spinal cord.

Peripherally, Elischer has described hyaline swelling of the axis cylinders of nerves along with increased interstitial tissue. Frey found changes similar to those of multiple neuritis.

H. M. Thomas in 1901 recorded 7 cases of embolism of the central artery of the retina associated with Chorea.

By far the most frequent lesion in Sydenham's Chorea is Endocarditis. According to Osler, with no disease, not excepting rheumatism, is it so constantly associated. Out of 73 autopsies of which he collected /

lected records, endocarditis was present in 62. Fagge found it in 17 out of 18 autopsies. The question then arises, Is the endocarditis in Chorea the result of Chorea or of previous Rheumatism? In 40 cases of Chorea with organic heart disease Sir W. Gowers attributed the endocarditis to previous Rheumatism in only 18. This estimate was probably too low.

A SHORT ACCOUNT of the BACTERIOLOGY of CHOREA.

BACTERIOLOGY.

Nannyn^u in 1888 reported the finding of a cladothrix on the meninges and the endocardium in a fatal case of Chorea.

Steinkopf found streptococci and Pianese a diplococcus and a staphylococcus in the nervous system. Leredde in 1891 found a streptococcus in the blood. Triboulet in 1893 found a staphylococcus in 2 cases at autopsy on one of which we had previously found a staphylococcus during life. In another case he found a diplococcus during life. Meyer reported finding streptococci and the staphylococcus citreus.

In /

In 1894 Dana found a Diplococcus in the case of Chorea complicated by chronic leptomeningitis already referred to. In a severe case of rheumatic endocarditis and chorea Triboulet and Ceyon discovered Achalme's Bacillus 40 hours after death. This is a large, motile, gram staining, anaerobic bacillus forming terminal spores. Its length varies according to the culture medium, being short when grown in media containing milk, sugar or glycerin and long when in ordinary ^{banilla} ~~banillas~~. It liquefied gelatin. Inoculation produces sero-sanguineous oedema at the point of inoculation and exudation into serous cavities. By injecting the pleural exudate of infected guinea-pigs into rabbits Thiroloix produced a condition resembling acute rheumatism especially in its cardiac manifestations.

Hewlett pointed out that Achalme's Bacillus is probably identical with B. Enteritidis Sporogenes of Klein.

In 2 cases of Chorea Apert (1898) found Triboulets Coccus or Diplococcus Rheumaticus, Westphal, Wasserman and Malkoff found diplococci in the brain of 1 case.

The researches and experiments of Poynton and Paine 1901, with the Diplococcus Rheumaticus and their confirmation by Beattie have been already referred /

referred to under Rheumatism as a factor in Etiology. D. Rheumatism is a minute coccus about .5 m in diameter. In the tissues it usually occurs in pairs ; in fluid cultures it forms short chains ; whilst on solid media it is irregularly arranged in masses. It stains readily with ordinary basic dyes and grains stain is not lost.

In a case of Cerebral Rheumatism, Souques and Castaigne found the Cerebro-spinal fluid sterile. Pic and Lesieur found Achalme's Bacillus in one case while in another cultures were sterile. Achalme himself found the cerebro-spinal fluid sterile although the blood contained anaerobic spore-bearing bacilla.

Staphylococci and streptococci have been found by different observers in the blood, cerebro-spinal fluid, brain and heart.

Blood.

Macalister has described experiments to determine the presence or absence of a toxin in the blood of patients suffering from Chorea. The result of these is to shew that (1) there appears to be in the blood plasma of Choreic patients a poison toxic to leucocytes of a healthy person. (2) In rheumatism the plasma is not so uniformly toxic. Macalister found that the blood corpuscles of a choreic /

choreic patient in the plasma of another choreic patient will live nearly as long as the blood corpuscles of a healthy person in the plasma of another healthy person, thus shewing that the blood cells become somewhat immune to the toxin. Therefore if the toxin of acute rheumatism is the same as that of Chorea the same immunity should exist when the blood corpuscles of a rheumatic patient are placed in the plasma of a choreic patient. When this was tried it was found that the life of the corpuscles was shortened. We may therefore infer that there is some difference between the poisons of rheumatism and Chorea.

In Chorea it is common to find eosinophilia to the extent of 16-20 per cent. This does not occur in Rheumatism. The eosinophilia remains for some time after the patient has apparently recovered from the attack of Chorea. These observations of Macalister tend to shew that there is less connection between rheumatism and chorea than is generally supposed, and that Chorea may be due to an infective toxæmia of distinct character. According to Murray Bligh, Von Pirquet's tuberculin reaction is always present in Chorea.

SYMPTOMS.

The symptoms of Chorea present all degrees of severity. They may be so slight as to be only perceptible, the so-called "Latent Chorea" of Miller. In mild cases the disease comes on insidiously. The first thing to attract the parent's notice is that the child is restless and cannot remain still, the condition popularly known as "fidgets". This begins in the arms or the face, less frequently in the legs. While sitting down the child will pronate and supinate the arm or open and shut her hand. When commencing in the face she will contract her facial muscles, drawing up the angle of the mouth, opening and shutting the eye, pursing of the lips, frowning, etc., etc. As the case progresses the movements spread to the trunk and the patient shrugs her shoulders and twists herself in a variety of contortions. Ultimately the movements reach the legs and the trunk is jerked from side to side. Walking becomes very difficult or impossible as she begins to lose control over her legs. The legs are rotated and thrown in front of one another. She becomes cross and irritable. As the disease gets worse the interval between the movements becomes shorter until ultimately the patient is in a state of constant movement only interrupted by sleep.

The movements are spontaneous and entirely involuntary /

voluntary. Later they become inco-ordinate and quite beyond control. There is loss of muscular power and this accounts for the want of precision in voluntary movements. Inco-ordination may be noticed before the spasms. The motor centres do not obey the will. When the patient tries to relax a set of muscles to complete some act there is delay. For instance when told to put out her tongue she takes a little time to open the mouth. Suddenly the mouth opens and the tongue is jerked out and as suddenly jerked back again as the jaws come together involuntarily with a snap. Or the centres may cause relaxation of a set of muscles during the performance of some act with disastrous results, e.g. dropping things on to the ground by involuntary relaxation of the muscles of the arm and hand. She may be unable to put her hand accurately upon an object.

Speech and deglutition may become seriously interfered with, the difficulty in the former being articulation. The speech may be reduced to a hopeless, meaningless jumble. Soon the patient can do nothing for herself. She can neither feed nor undress herself. She may become so bad as to be unable to lie in bed, one of my patients requiring considerable mechanical restraint to prevent her injuring herself.

In many cases muscular weakness is the prominent feature. It is usually only of the nature of paresis, but /

but it may develop into paralysis. When it is a paresis it is usually exhibited by a weakness of handgrip or dragging of a leg. Muscular weakness is most marked in cases with little spasm. It is often of a hemiplegic type and called Chorea Mollis by C. West, and Paralytic Chorea by Sir W. Gowers.

The muscular weakness may come on in cases already spasmodic or the weakness may appear before the spasmodic movements or after they have ceased. When they are present together they usually disappear together but sometimes the paresis persists after the spasms have gone. Osler reports a case where wrist drop persisted in the right arm for 2 years after Chorea. Dr Judson Bury reported a case of foot-drop, persisting 4 years after the onset of paresis.

Von. Ziemssen has observed irregular movements of the vocal cords but it is doubtful if the contractions spread to the muscles of internal organs. Movements of the thorax in respiration may be disorderedly owing to implication of the diaphragm. They become irregular in time and depth. This may cause irregularity of the heart quite apart from organic disease.

The movements of Chorea are arrested by sleep. Dr J. W. Russell pointed out that they may sometimes be inhibited by making the child attempt to write. Russell, Weir Mitchell, and Rhein divided their cases according to the character of the handwriting.

Chorea may be unilateral with regard to limbs but when affecting the face and trunk it is always bilateral. (Hughlings Jackson.) The left side is affected slightly more often than the right. (Pye Smith and Gowers.) All my cases have been bilateral.

Electrical Reactions.

These have been specially studied by Benedikt, Rosenthal, Schmidt, and Gowers. These observers agree that in some cases the reaction of the muscles to the faradic and galvanic currents is increased. In some cases the anodal closing contraction has been equal to the kathodal closing contraction instead of being weaker.

In none of my cases was I able to detect "Reaction of Degeneration."

Heart Symptoms.

Tachycardia is very common and often due to the fact that the patient is a neurotic girl. As already mentioned, it may be due to irregular movements of the thoracic muscles. It has been ascribed to Chorea of the heart but this is doubtful. It is usually regular and not associated with any praecordial pain.

Murmurs may be heard both at the apex and the base. These may be either due to organic disease or to /

to anaemia and it is important with regard to diagnosis and prognosis to make a careful distinction. According to Risien Russell doubt can only arise in the case of a systolic murmur ; presystolic or early diastolic can only mean organic disease. Others maintain that a presystolic or early diastolic murmur may be haemic in character. The most common organic murmur is mitral systolic. Aortic organic disease is very rare in Chorea. The Collective Investigation Committee of the British Medical Association found 116 cases of mitral organic disease and 6 or aortic organic disease, In 250 cases examined by Sir W. Gowers only 2 had aortic organic disease.

Organic murmurs are usually heard at the apex and haemic murmurs have their maximum intensity at the base. An organic mitral systolic murmur is propagated into the axilla but even here difficulty in diagnosis may arise for a murmur propagated into the axilla may be due to anaemic dilatation of the left ventricle. One must clear up the diagnosis by looking for other signs of anaemia, e.g. venous hum in the neck, etc. Haemic murmurs at the apex are not usually propagated beyond the praecordia. Hypertrophy of the heart wall will also be found along with organic disease. Endocarditis and anaemia may both be present. In such cases the curing of the anaemia does not give actual proof of the nature of /

of a murmur for an endocarditic murmur may disappear at the same time.

The endocarditis is usually of a simple nature but it is likely to cause sclerosis of the valve or valves affected. Osler examined 140 patients two years after the attack of Chorea and found organic heart disease in rather over 50 per cent. According to Gowers in 9 out of every 10 fatal cases of Chorea valvular disease is found. Out of 80 cases only 5 had the heart and pericardium free from disease. (Sturges.) Of my own cases, 46 in number, endocarditis was found in 39.

An organic murmur may be present before the attack of Chorea, due to previous Chorea or acute rheumatism. First attacks of Chorea may be developed without any murmur supervening until the patient has quite recovered from the Chorea.

Pericarditis occurs occasionally but usually along with severe rheumatism and endocarditis. Sibson recorded 21 cases of Chorea associated with acute rheumatism, 15 developed pericarditis, 14 endocarditis, 3 doubtful endocarditis, 6 no pericarditis, 3 no endocarditis. In other 73 necropsies 19 were found to have pericarditis in only 2 of which endocarditis was absent.

Sensory Symptoms.

Pain in the affected parts is not common and is usually in cases of hemichorea. There may be headache either continuous or paroxysmal. There are sometimes painful points along the spine at the exit of spinal nerves from the deeper structures (Cartier, Triboulet and Marie.) Trousseau occasionally found tingling and formication in affected regions. Impairment of sensibility is probably hysterical in character. Blunting of sensibility is common according to Fere and Broadbent. Purves Stewart found it in 10 out of 41 cases. Of my own cases continuous headache occurred in 8, paroxysmal in the majority. In 6 there was considerable impairment of sensibility but in none was there any acute pain.

Reflexes both superficial and deep are usually unaltered, but they may be exaggerated, diminished, or even in paralytic cases abolished. Sinkler found that of 50 cases the knee jerks were normal in 26, diminished in 15, absent in 9. Oddo found that of 147 cases the knee jerks were normal in 13 per cent, increased in 25 per cent, diminished in 62 per cent. According to Gordon the knee jerk is altered peculiarly. The limb is suspended in the air too long after the patellar tendon is struck owing to prolonged contraction of the quadriceps.

This /

This has been explained by Dr Judson Bury who declares that Choreic movement can be excited by percussion of a muscle or tendon.

Trophic changes in the affected muscles are very uncommon.

Sphincters are usually not affected although there may be involuntary evacuations in very severe cases with the possible development of bed sores.

Psychical and Mental Symptoms.

These have been most studied by Marce, Buccelli, and Burr. They are usually very well marked. The patient is usually neurotic and often the child of neurotic parents. She is usually wilful and bad tempered and gives way to violent emotional outbursts of tears or temper on the slightest provocation.

The degree of mental change is not relative to the severity of the other symptoms. The memory is usually enfeebled, and the power of concentration of the mind lessened.

According to Axenfeld and Huchard there may be hallucinations. These are usually of sight or some other special sense and very rarely of the sexual sense. Edes has described acute melancholia associated with Chorea. This may partake of a suicidal character. There may be acute mania or Chorea /

Chorea Insanieus. Delirium is usually recovered from but it may be the starting point of permanent mental deterioration in cases with a hereditary taint of insanity.

Burr insists that there are no mental symptoms of Chorea and no such disease as Chorea Insanieus.

Convulsions are very rare.

Temperature may be raised but this is usually due to acute rheumatism or to endocarditis. According to Hare in hemichorea the temperature of the affected side may be raised but this is not by any means invariable. Hyperpyrexia without complications is excessively rare but G. Carpenter records a case which terminated fatally.

Skin.

Associated with Chorea various rheumatic eruptions occur occasionally, e.g. Erythema nodosum, purpura^urticaria, peliosis rheumatica. More rarely are found subcutaneous fibrous nodules and Herpes Zoster. Care must be taken to distinguish a rheumatic skin eruption from one due to arsenic with which the patient may be being treated. Alopecia areata is said to have occurred but the bald patches may be due to constant rubbing of the head on the pillow.

Urine /

Urine.

Various abnormal constituents of urine have been found. Walshe found an excess of urea probably due to the abnormal muscular activity and increased metabolisms. Todd, Herter, and Smith found a considerable deposit of uric acid due to disturbance of nutrition. Handfield Jones found a large increase of phosphates. These are proportionate to the severity of the Chorea. Urohaemato porphyrin was found by Dr A. E. Garrod in 14 out of 20 cases, this being additional evidence of the relation of Chorea to Rheumatism. Albumin when present is usually due to co-existing nephritis. Glycosuria has been reported as occurring occasionally.

Ocular Symptoms.

Strabismus frequently occurs due to spasmodic contraction of the extrinsic muscles of the eye. The pupils are often dilated especially on the side with the most severe spasms.

According to Langmead the following alterations in the pupil are fairly common (1) Hippus. This is a condition in which the iris has a wide extent of movement. The movements are very rapid owing to the jerking of the eyeballs, the frequent variation in the amount of light reaching the retina, and the sudden altering of the accommodation. This sometimes /

times happens during intervals of quiescence.

Langmead says that hippus is not often met with except in articular and cardiac rheumatism. Dr

R. W. Phillip has reported a case of Chorea with

hippus. (2) Peruliar movements of accommodation.

Contraction to accommodation is very rapid and may be asynchronous. Contraction both to light

and accommodation is usually ill-sustained and may also be asynchronous. (3) One pupil may remain

smaller than the other for a considerable time. (4)

Eccentricity of the pupil. This very often is best

marked when the pupils are contracted. Miller

regards these anomalies purely as signs of nervous instability.

The optic discs are usually not affected.

There is occasionally neuritis, sometimes put down to co-existing hypermetropia but it may disappear without

correction of the error of refraction. It is more

probably due to some infective state of the blood

than secondary to an intra cranial lesion.

DIAGNOSIS /

DIAGNOSIS.

The diagnosis of Sydenham's Chorea is usually very easy and may be made at a glance. The irregular, spontaneous, purposeless movements associated with inco-ordination are as a rule quite typical of the disease. Sometimes, however, there may be difficulty experienced particularly in the cases with paralytic symptoms. Paralytic Chorea without movements may be very difficult to diagnose from other forms of paralysis. The weakness in Chorea usually comes on gradually, but it may be sudden in onset. The diagnosis becomes much more difficult when hemiplegia is associated with endocarditis. In such cases there is no history of convulsions nor of loss of consciousness ; there is no paralysis of the face ; and the paralysis of the limbs is not spastic as in occlusion of a cerebral vessel.

In monoplegic weakness, e.g., of one arm - the most common form of paresis in Chorea - again the slow onset distinguishes Chorea from an organic affection. Chorea may be confounded with cerebral disease in infancy, but the history of recent onset helps towards the diagnosis. Anterior poliomyelitis may be mistaken for Chorea in cases where the Choreic movements are slight and the paresis the dominating symptom. The absence of atrophy of the muscles and of the reaction of degeneration help to clinch the diagnosis in favour of Chorea.

Peripheral neuritis is distinguished by the sensory symptoms and the reaction of degeneration, while Chorea may be diagnosed by perhaps the detection of slight spontaneous movements, inability to tightly grasp an object for a considerable time, or delay in relaxing some group of muscles.

Multiple diffuse cerebral sclerosis (sometimes called Chorea Spastica) is characterised by exaggeration of the knee jerks and the tremor is more rhythmical-nystagmus also present. Marked paresis of both legs may simulate a paraplegia of spinal origin.

Paralysis Agitans. rarely occurs under the age of 40, but cases have been reported as occurring at 20. Here the tremor is much finer and we have the characteristic mask, attitude, and gait.

Friedreich's Ataxia is distinguished by its marked hereditary characteristics and by slow irregular, inco-ordinate movements, nystagmus, scanning speech, and talipes.

Paramyoclonus Multiplex. In this disease the spasms are more sudden and shock like. It affects similar muscles on both sides, and is commonly confined to the trunk and the proximal muscles of the limbs. It is more persistent than Chorea although more harmless. It has no connection whatever with rheumatism or endocarditis.

Athetosis is characterised by slow undulating movements, and stiff limbs. The muscles are often increased in size. Convulsions frequently occur, and defective intellect is common.

Huntington's Chorea. This rarely develops before the age of 40. It has a very marked hereditary character. It consists of irregular movements beginning in the hands and spreading to other parts of the body. The movements are not so sharp as in Sydenham's Chorea, and the gait is swaying. Speech is slow and difficult. It persists until the end of life, and is accompanied by marked mental deterioration.

Electric Chorea (Dubini's Disease). This condition is only found in Italy. Spasms are very sudden and shock like, and give their name to the disease. The course is progressive. There are epileptiform convulsions, paresis, wasting of muscles, and loss of Faradic response. Death often occurs with hyperpyrexia.

Hysteria. The diagnosis between Chorea and Hysteria may be very difficult and it is probable that the cases of Chorea which have been reported as due to imitation are really not Chorea at all, but a purely hysterical manifestation. The patients are usually girls at the age of puberty.

Acute Mania must be distinguished from the mania associated with chorea in which the choreic movements may usually be discerned. These cases are sometimes not diagnosed till the patient has been sent to an asylum where the true nature of the disease is found.

Habit Spasm. Chorea must be carefully distinguished from the habit spasm which is sometimes found in neurotic girls of 8 to 14 years of age. Habit spasm consists of quick contraction of the facial muscles e.g. drawing the mouth to one side, winking, etc. Neck muscles may be involved and shrugging of the shoulders. These movements are repeated at irregular intervals and the condition closely simulates Chorea.

Impulsive Tic. This is a hysterical condition affecting young children of neurotic family and characterised by involuntary muscular movements usually of the face and arms but occasionally of the whole body and by explosive utterances resembling a bark. Word mimicry or echolalia and action mimicry or echokinesis also occur. The movements have a wider range than those of chorea and are of an explosive nature.

DURATION and RECURRENCE.

An ordinary moderate case of chorea usually runs a course of from 6 to 10 weeks and ends in a perfect recovery. A few cases run a protracted course of several months. Two of my cases were cured after being under treatment for 7 and 8 days respectively but they were very mild cases. The most protracted case was that of a girl aged 9 who was under treatment in hospital for four months before being discharged cured. Between these extremes were the others. The average stay in hospital was 60 days. One attack predisposes to another, a fact which was recognised by Sydenham. A second attack may occur after a few months interval. The average interval between first and second attacks is about one year. If only a few weeks elapse, the second attack is more likely to be a relapse of the previous attack than a recurrence. There may be several recurrences. Of 410 cases, 240 had one attack, 110 had 2 attacks, 35 had 3 attacks, 10 had 4 attacks, 12 had 5 attacks, and 3 had 6 attacks. Gowers reports a case of 9 attacks. Recurrence usually takes place in the Spring. As the heart is more frequently affected in a recurrence than in a primary attack, some say that cardiac disease favours recurrence. The reverse is more likely i.e. the greater the number of attacks, the more likely is endocarditis to develop. Endocarditis /

ditis has been found in a recurrence when there was no trace of it in the primary attack.

Just as to a primary attack, females are more prone to recurrence and are more likely to have several attacks.

Recurrence has been ascribed to fright, but in many cases no evidence of fright can be got. Overwork at school may be an important factor in causing a recurrence. Pregnancy may cause a second or third attack, but very rarely more.

The symptoms in a recurrence are not usually so severe as in the primary attack but are occasionally more so. Usually the same regions are affected.

Prognosis. /

PROGNOSIS.

The prognosis in the great majority of cases is good. Recovery is usual in children even in severe cases. The mortality is about 2 per cent. The Collective Investigation Committee found 9 deaths in 439 cases or 2.05 per cent. 64 deaths occurred in Philadelphia in 74 years (Sinkler.) I myself had 1 death in 46 cases or 2.17 per cent. Death in a recurrence is rare. The mortality increases with the age and is high in the chorea of pregnancy. (Anstie) French and Hicks deny this. With them only 3 out of 29 cases of Chorea Gravidarum died and in these fatal cases the temperature rose above 100° F without any complication arising. Most deaths in Chorea gravidarum occur from abortion either spontaneous or induced. Full term labour may prove fatal from the extreme debility of the patient. But H. K. Wallace reports a case where Chorea developed in two successive pregnancies. In the first pregnancy the choreic movements began a week before labour and in the second a month before labour. Delivery with forceps on both occasions and a good recovery. The patient had suffered from Chorea at the age of 14 years

The degree of psychical disturbance is no indication for a fatal prognosis, although permanent mental deterioration /

deterioration may follow. Mental effects may last for years and then disappear, and epileptiform attacks may develop into true epilepsy.

Apart from complications the chief danger lies in exhaustion from the never-ending violent movements, in mal nutrition, and in want of sleep. A first attack usually lasts longer than a subsequent one. The more severe the spasmodic movements, the more likely is the attack to last a long time. The paralytic cases are the most obstinate but Choreic paralysis usually disappears.

The hysterical form of Chorea is never fatal.

Endocarditis does not affect the immediate prognosis but it has a great influence over the ultimate progress of the case. Pre-existing endocarditis is aggravated by attacks of Chorea, leading to structural changes, failure of compensation, etc.

Forssmer followed up 28 cases treated at Stockholm from 1885 to 1892. Seven died of heart disease ; 7 had severe heart affection ; 5 developed phthisis ; 7 albuminuria along with heart disease and 1 of these had a goitre ; 1 had goitre with heart disease, probably Basedow's disease ; 1 had some gastric disorder ; 1 was mentally affected. Only 1 out of the 28 was in good health. Five of the cases with heart disease had died before reaching the age of 11 years. Of the remaining 23, 14 had some chronic affection not a complication of Chorea.

TREATMENT.

Prophylaxis. A certain degree of precaution is possible in order to prevent an attack of Chorea. Bright intelligent children of neurotic parents should be carefully watched during their school days and should never be encouraged to work hard for school honours. Plenty of opportunity should be given for open air and exercise and the brain should never be overtaxed with lessons.

Treatment of the Attack. The first thing to do is to find the exciting cause if possible and to remove it. All reflex sources of irritation such as adenoids, errors of refraction, constipation, worms, phimosis, etc. should be attended to. In Chorea Gravidarum consider emptying the uterus. French and Hicks say that procuring abortion is no cure, that the case should be treated on the same lines as any other case of Chorea, that most cases get well without it, and that it is too late to induce labour after the temperature has begun to rise. In H. K. Wallace's case the drugs in common use in Chorea had little effect. E. O. Croft of Leeds declares that induction of labour is wrong treatment as it does not stop the Chorea. The case should be treated on general principles and the pregnancy allowed to go to full time. W. F. Shaw of Manchester is of the same opinion.

In the treatment of very mild cases of Chorea all that may be necessary may be to stop school work and all mental effort.

In a more severe case general hygienic treatment is the first necessity. The child should be kept in bed and in absolute quiet. No other children should be allowed into the room. If in hospital it is well to have the bed surrounded with screens. There is thus a minimum of excitement and this tends to allay the nervous excitability and to prevent damage to the valves of the heart should endocarditis be present. If in a private house, and especially where the mother is neurotic, a sensible trained nurse accustomed to children is an advantage. The child should be kept in bed till the choreic movements cease. Constant rest in bed may be irksome and do harm by causing mental depression. In such cases the child should go to bed early and get up late and she should lie down for a few hours in the afternoon.

The diet should be nutritious and easily digested. All stimulants, e.g. tea, coffee are absolutely forbidden. The bowels should be carefully regulated and constipation avoided.

In very bad cases the child must be protected from injury. The mattress should be soft. If obtainable a water bed is a distinct asset. Bed sores should be carefully guarded against. All prominent /

prominent bony points should be protected. It is a good plan to tie a pillow to the inside of one leg. It may be necessary to use mechanical restraint to prevent the patient jumping out of bed. In such cases the bed may be conveniently placed on the floor.

Warm baths are good if the spasms are not too severe. The cold pack may be employed if the temperature is high. ^{very} ~~Pastes~~ strongly recommended the hot pack.

Which?

Medicinal Treatment.

A large number of drugs have been tried, singly and in combination, in the treatment of Chorea. The obvious inference is that there is no particular one which acts as a specific on the disease. Some prefer one drug, others another. I shall endeavour to give an account of those in most general use, with the results obtained with each, and also of a few which have been tried experimentally, but have not yet found their way into ordinary practice.

The drug in most common use and really the only one which appears to have much effect is Arsenic. Its specific action is not known. It probably has none but only produced its effect by improving the general nutrition of the patient. It is usually prescribed as Fowler's Solution (Liquor Arsenicalis).

The /



The dose varies. Some physicians (and I number myself amongst them) give large doses commencing with 10 or 15 minims. I usually prescribe for a child of 10-12 years 10 minims thrice daily, after food in a large quantity of water. I increase the dose by 1 minim every alternate day carefully watching for any sign of intolerance until the patient is taking 15-20 minims thrice daily. The danger lies in causing arsenical poisoning, but I have found that children with Chorea tolerate these large doses very well. When toxic symptoms begin to shew, the drug should be stopped for four or five days and then resumed with a slightly smaller dose than that which was being taken when the stoppage occurred. Cases of intractable neuritis have been reported as resulting from these large doses of arsenic, but I have not seen one. Cary Gamble reports a case of fatal arsenical neuritis in a child aged 8 years who took m. vii. t.d.s. for 10 days, stopped the drug for a week, and then resumed m. vii. t.d.s. for 14 days.

Many physicians declare that these large doses of arsenic are quite unnecessary, and that equally good results are obtained with small doses of m. iii. - m. v. Guthrie Rankin said that arsenic was the best drug for Chorea, and should be given in doses of m. iii. increased to m. x. Koplik objects to big doses on account of danger to the Kidneys.

Eulenburg, Hammond, and Weiderhofer recommended that arsenic should be given subcutaneously instead of by the mouth. They claimed that larger doses could be given without causing intolerance of the gastro-intestinal tract and that even when intolerance was established the drug could be continued. It is probable that such treatment would set up neuritis. The plan is feasible in adults but obviously not in children. Cacodylate of Soda has been much used in France instead of Fowler's solution. It may be given hypodermically, or as a pill in larger doses than other preparations of arsenic. Chapin shewed experimentally that arsenic is less toxic when combined with butter than when given in solution. A known quantity of arsenious acid according to the dose to be given is added to Sodium Chloride in the proportion of .005 gram to .1 gram. The mixture is triturated with 10 grams of fresh butter and given spread on bread after meals. If the patient is anaemic, iron may be combined with arsenic or may even be substituted. In weakly children, the Syrup of the Phosphate of Iron along with Cod Liver Oil, Phosphate of Lime, Malt, etc., are useful adjuvants or substitutes for arsenic. Teubert strongly recommends arsentriferrin (a mixture of arsenⁱⁿparanuncⁱⁿclⁱⁿimate of iron and paranuncⁱⁿclⁱⁿimate of iron) /

iron) as a combination of iron and arsenic in the treatment of Chorea. In definitely rheumatic cases, salicylate of soda may be given in the early stages.

D. B. Lees prescribed this drug along, with twice the dose bicarbonate of soda to aid toleration, if there are any cardiac symptoms or any acute rheumatism present. Leonard Guthrie maintained that large doses of Salicylate of Soda were not without danger, and that the combination with bicarbonate of Soda was questionable therapeutics

personally, I have known that Aceto-Salicylic acid (aspirin) is better than sodium salicylate in rheumatic cases in doses of 10-15 grains three or four times daily, and my experience is corroborated by that of Williamson. Dr Cecil Wall recommended aspirin as the best drug for Chorea. His results were that 82 per cent of cases left hospital in less than two months - the majority in four weeks - when treated with aspirin and only 38 per cent when salicylate of soda and other drugs were used. Aspirin should be given in powder form (not in tablets) on a full stomach.

Lees reports a case of coma produced by aspirin. There is less likelihood of cardiac depression with this drug than with Salicylate of Soda. Very large doses will ^{cause} ~~call~~ haematuria (Wall.)

H. /

H. C. Wood has obtained good results by giving large doses of Quinine. This drug is such used in America. The Sulphate and the Oxide of Zinc in doses up to 20 grains have been tried ; also the valerianate and bromo valerianate but without any very convincing results. Strychnine is not to be recommended in the early acute stage, but it is undoubtedly useful in the later and in the paralytic stages.

Of the sedative drugs, the best undoubtedly is Choral. Good results were got by Sir W. Gairdner and by Charlton Bastian. The patient was kept continuously under the influence of the drug in order to procure arrest of the choreic movements by prolonged sleep. This form of treatment should only be adopted in very intractable cases and a careful lookout must be kept for cardiac depression and gastric irritation. In some cases the natural course of the disease is not shortened, and in others the spasmodic movements return when the drug is stopped. There is also the danger of causing mental derangement. Bromides are of little use unless in combination with Choral when some benefit may certainly accrue. In severe cases, R. W. Phillip believes in giving doses of 30 grains of each drug. Good results have occasionally been obtained in Chorea Gravidarum. Camphor monobromate has been used by Ghetti in doses of $\frac{3}{4}$ grain in a cachet thrice daily for a week, and four times daily for another week.

He thought that recovery with this drug was more rapid than with arsenic. Bourneville and Katz report rapid cure with bromide of camphor in capsules of 20 centigrams. The hydrochlorate and hydrobromate of hyos@ine are sometimes useful in severe cases. Babinski has used the hydrobromate of scopolamine. The latest preparation of bromide in use for Chorea is Sabromin. According to Maetzke it is less likely to cause bromism than the ordinary bromide salts. It is a combination of bromide with calcium and is prescribed in tablet form, 3 to 6 tablets (25-45 grains) to be given daily.

Other sedative drugs in fairly common use are Sulphonal, Trional, Paraldehyde, and Veronal. Chloretone or Trichlor-tertiary-butyl-alcohol ($C_4H_7OCL_3$) has been strongly advocated especially by Wynter. It is prescribed in doses of 5 grains in half a drachm of petroleum emulsion every 4, 6, or 8 hours, according to the severity of the case. As the disease improves the daily quantity of the drug is diminished. In 50 cases treated with Chloretone the duration of treatment averaged 9 days. Absorption is accelerated, and the action of the drug assisted by gentle laxatives. Excessive doses will produce stupor, erythema, and peeling of the skin of the hands and feet.

Antipyrin in doses of 2 drachms in 24 hours may
do /

do good in the early acute stage, but albuminuria and other signs of intolerance must be watched for.

Exalgin, Analgen, Asaprol, Physostigmine, and even Curare have been used, the last named is very dangerous. Opium is given to children in large doses by Jaccond. Cimicifuga was used with great success by ^{M. J. Merney} ~~McMerney~~. The patient was a child aged 10 years with acute rheumatism and severe Chorea with loss of speech. The rheumatism disappeared with appropriate treatment, but, the Chorea persisted. The liquid extract of cimicifuga in doses of m xx. every four hours was given. The power of speech returned in 10 days, and the child was perfectly cured of the Chorea.

Conium juice in large doses is given by Ringer, but it upsets digestion and the effects are not permanent.

Corrosive sublimate. Riva working on the hypothesis of a bacterial causation of Chorea tried intravenous injection of this drug in doses of one/twentieth to one/sixth grain, and found that it was well tolerated by children.

Marinescu of Bucharest describes four cases treated by intraspinal injection of Magnesium Sulphate. He used a 25 per cent solution of the pure crystallized salt, which must be freshly prepared. The dose given was 1 cc. of the solution for each

25 pounds of the patient's weight. In two cases there were striking results. One was completely cured in three days with one injection, and the other in seven days with two injections. The injection caused intense headache (which was relieved by morphine) tingling, numbness, inability to move the legs, and nausea.

Roden reports a case of a child of 10 years with Chorea. The heart was sound. All the ordinary drugs failed. A temporary cure was effected with brine baths. The same result happened with a second attack. While attending a third attack he noticed that the patient's two sisters had goitre. He put the patient on Fluid Extract of Thyroid Gland (Duncan & Flockhart), in doses of 1 drachm twice daily. The child returned to school in the second week of treatment, which was continued for a month. In the second month the dose was reduced to once daily. At the time of writing (three years later), there had been no further recurrence.

Salvarsan has been successfully used by Mayerhofer in a case of post-rheumatic Chorea minor which had persisted for nearly three years in spite of all ordinary methods of treatment. Wasserman's reaction was negative ; Von Pirquet's positive. An intra^{mus}cular injection of 0.25 gram hyperideal in acid solution was given with improvement in the symptoms /

symptoms. A month later 0.5 gram was given. Within six days of the second injection the Chorea had disappeared. A similarly successful case had been reported by Bokey.

Dr Ainley Walker has published an interesting paper on the cure of rheumatism by Bee-stings and mentions Dr Terc, an Austrian Physician of Marburg (Styria) who had carried out the treatment for years. Dr Muir Evans drew attention to a phenomenon described by Phisalix in 1904. This observer found that Bee-stings produced Chorea form movements in sparrows. Dr Muir suggested that small repeated doses of the poison might elaborate antibodies and thus confer immunity to rheumatism and chorea. The idea is certainly grotesque.

At the conclusion of this account of the drug treatment of Chorea, I should like to quote "D'Orsay Heck and Wall who deprecate the use of drugs altogether and maintain that the patient recovers in the same time with general hygienic treatment and no drugs.

Vaccine treatment :-

R. J. M. Buchannan has quite lately described a vaccine treatment for Chorea. He concluded that the diplococcus of Poynton and Paine was the cause. He made a vaccine from streptococci obtained from the blood of a patient suffering from rheumatic endocarditis /

docarditis. He reports that he has obtained good results with this in Chorea, but experience of the treatment is yet too limited to allow of a definite value being put upon it.

The treatment of special symptoms must be attended to. In severe cases sleeplessness is very obstinate owing to the constant movement. For this Chloral, bromide and the other hypnotics, including morphia, if necessary, may be given. Very often all efforts to induce sleep fail, and the patient sinks into a state of exhaustion with rapid heart failure and death. Since cases may require cardiac stimulation from the beginning, but not, I should think, with very good results.

When recovery is taking place and the choreic movements have practically ceased, a little Massage of the muscles is of benefit. This is especially useful in the paralytic forms.

Electricity in the form of a weak galvanic current may be applied in the late stages, and in paralytic cases. Faradism is not to be recommended.

A system of exercises has been planned by Guthrie based upon Frankel's system for Locomotor Ataxia and having for their object the re-education of the patient in voluntary movements. These exercises have been divided into four stages, and may be briefly described.

1. The child should be persuaded to lie in bed as quietly as possible, and the movements should be gently restrained.
2. This is the stage of passive movements combined with suggestion. Choreic movements will occur whenever any attempt is made to manipulate a limb. The patient's hand should be held between the operator's hands and moved in various directions, the child being told to keep her hand as still as possible. At first she will snatch it away, but in course of time she acquires a certain amount of control, and holds the limb rigid. It is a sign of improvement when the efforts to secure control require to be less strenuous and the rigidity becomes relaxed. Until the arm can be moved passively in all directions without spasm or rigidity, only passive exercises should be employed.
3. Voluntary movements under guidance. The child is told to perform some movement, which has been previously performed passively, the limb being at the same time guided and controlled.
4. When the third stage can be gone through without spasms or rigidity, the patient may then attempt voluntary movements without control.

All movements must be simple at first and gradually /

ually elaborated. The chief difficulty is inco-ordination. Such exercises as writing, sewing, or threading needles should not be allowed as they require too much mental concentration. All exercises should be done under careful supervision and the child not allowed to get careless or she will drift into a state which might be termed "residual Chorea."

Exercises for the legs should be conducted on the same principle as those for the arms.

The principles of these exercises are (1) that suggestion is of service where there is lack of inhibitory control over the Choreic movements. (2) that inhibition when acquired is often exaggerated and has to be regulated by assistance before voluntary movements can be executed. (3) that inco-ordination has to be treated by exercises carefully graduated in the order of the difficulty of their accomplishment.

After recovery from the attack the child should not be sent to school for a week or two and the return to school work must be gradual, the effect being watched very carefully. On the reappearance of the slightest sign of choreic movement all work must be immediately stopped.

Change of air and scene is beneficial in the convalescent stage and often works great improvement in those cases which drag on for months under treatment without much apparent result.

CONCLUSIONS.

I have in these pages set forth the result of my study of 46 cases of Sydenham's Chorea aided by reference to the literature of the subject embodying the experience of others. I shall now briefly summarise the conclusions at which I have arrived.

These are :-

1. That, with the exception of the Chorea of Pregnancy, Chorea is a disease of childhood.
2. That there is a large preponderance of females over males.
3. That Chorea is a late manifestation of some infectious agent which in a considerable proportion of cases is Rheumatism. After analysing the researches of Poynton, Paine, and Holmes on the bacteriology of the disease and the experiments of Macalister with the blood, I am not convinced that the *Diplococcus Rheumaticus* is entirely responsible for the disease although there is no doubt but that in many cases it is.
4. That it is quite possible that the Embolic Theory of Kirkes may in some cases be correct. Endocarditis is a very common but not a universal concomitant of Chorea.
5. ,That Chorea due to imitation does not exist, all such cases being hysterical in character.
6. /

6. That the best drug to employ is arsenic (given as Fowler's solution) in large doses, commencing with 10 minims. In most cases the disease improved under its influence.

BIBLIOGRAPHY.(in alphabetical order.)

1. Babinski. Rev. Neurol, Paris, 1907 xv. 86.
2. Bastian, Charlton. Lancet 1889, ii, 55.
3. Batten. Lancet 1898 ii, 1195.
4. Buist. Trans. Edin. Obst. Soc. 1895.
5. Bulloch. System of Medicine by Allbutt and Rolleston.
6. Bury, Judson. Med. Chron. 1909, 61, 141.
7. Collective Investigation Committee of the British Medical Association.
8. Croft. Brit. Med. Journ. 1910 i, 872.
9. Dana. Amer. Journ. Med. Sc. Phila. 1894, cvii, 31.
10. Dickinson. Wilk's Nervous Diseases.
11. Evans, Muir Brit. Med. Journ. 1908, ii, 1409.
12. Forssner. Hospital July 2, 1910.
13. Gairdner. Lancet 1889, ii. 205.
14. Ghetti. Gaz. deg. Osped. Mar. 10, 1910.
15. Gordon. Brit. Med. Journ. 1901 i, 765.
16. Guthrie. Leonard. Brit. Med. Journ. 1908, 1, 1234.
17. Hewlett. Lancet 1901, i, 704.
18. Holt. Diseases of Infancy and Childhood.
19. Langmead. Lancet, 1908, i, 154.
20. Lees. Brit. Journ. Child. Dis. 1909 vi., 97.
21. Lewis Morris, Trans. Assoc. Amer. Phys. 1892, vii. 249.
22. Macalister, Brit. Med. Journ, 1909, ii. 514.
23. McCarthy. System of Medicine by Osler and McCrae.
24. /

24. McMerney ~~McMerney~~. Brit. Med. Journ. 1907, ii. 1380.
25. Maetake. Dent. Med. Woch, July 28, 1910.
26. Marinescu ~~Marinescu~~. Sem. Med. 1908, 553.
27. Mayerhofer. Wein. Klm. Woch, 1911, 27.
28. Miller, Lancet 1909, ii, 1808.
29. Osler. Principles and Practice of Medicine
30. Do. Chorea and Choreiform affections.
31. Pasteur Med. Review July 1910.
32. Phillip ~~R~~ W. Brit. Med. Journ, 1908, i, 365.
33. Poynton and Paine Lancet 1901, i, 1260
34. Poynton and Holmes, Lancet 1906, ii, 982.
35. Rankin, Guthrie. Brit. Med. Journ, 1908 ii, 699.
36. Roden, Lancet. 1910 ii. 1276.
37. Russell, Risien, System of Medicine by Allbutt and Rolleston.
38. Shaw, Lancet 1906, ii, 1196
39. Sinkler. Nerv. Dis. by Amer. Authors.
40. Teubert Berlin Klin. Woch July, 11, 1910.
41. Thirolloix Ibid, 1897, 882.
42. Townsend. Trans. Amer. Paediat Soc. 1892, 4.
43. Turner, Trans Path. Soc. Lond. 1892 xiii, 8
44. Walker, Ainley Brit. Med. Journ 1908, ii, 1056
45. Wall Cecil, Trans. Therap. Soc. 1905.
46. Wallace, Brit. Med. Journ, 1907, i, 260.
47. Williamson Lancet 1903 ii, 526.
48. Wood, Trans. Assoc. Amer. Phys. Phila, 1893.
49. Wynter, Essex Brit. Med. Journ, 1908, ii, 912.

FACULTY OF MEDICINE.

University New Buildings.
Edinburgh 2nd May 1902

Professor Greenfield

Dear Sir,

The accompanying
thesis by G. H. Lowe, M.B.
is referred to you for second
reading by Professor Wyllie.

Yours faithfully,

F. Harvey Littlejohn
Dean J.B.

having a Confusion - and a diff
to get much of this aspect - i.e. not
taken for other papers. That must have
involved a good deal of work.

Looks upon the numerous cases he has seen
that he gives no minds of them - & there is
a better aspect.

And he has not found much criticism
of the heterogeneous ~~the~~ mass of conflicting
opinions -

As to the amount of material the L.H. requires
whether by recommendation or otherwise
is doubtful. It depends on this.

My recommendation is all of the present

W.A.G.

